

given routinely at birth and she was fully breast fed, and was thriving and developing normally. At age 6 weeks she developed a blood-stained nasal discharge and 48 hours later became lethargic, reluctant to feed, and was noticed to be jaundiced. Six hours later she collapsed and had a grand mal convulsion.

When examined she was apyrexial, pale, and jaundiced with a firm liver edge palpable 3 cm below the right costal margin; she was unresponsive and hypertonic with a tense fontanelle. There was excessive bleeding from venepuncture sites and cerebrospinal fluid obtained from lumbar puncture was grossly blood stained. The haemoglobin concentration was 9.8 g/dl with a normal blood film; platelet count was normal but prothrombin time was more than 120 seconds.

Vitamin K and fresh frozen plasma were given. Within 6 hours her haemoglobin concentration fell to 6.8 g/dl. Liver function tests were abnormal (bilirubin 132 μ mol/l, unconjugated 80 μ mol/l, with increased levels of transaminases) but clotting tests were nearly normal (prothrombin time 19 seconds, control 14 seconds). A provisional diagnosis of acute encephalopathy was made and she was treated intensively with endotracheal intubation, hyperventilation, and high dose phenobarbitone in an attempt to reduce intracranial pressure. Ultrasound examination showed normal ventricles with no evidence of intraventricular haemorrhage but subdural effusions could not be excluded and as the fontanelle was still tense 28 hours after the initial seizure, bilateral subdural taps were done which showed heavily blood-stained cerebrospinal fluid under pressure on the right side.

There was no subsequent evidence of bacterial or viral infections. Six weeks after the acute illness she had begun to smile again but was generally hypertonic. There had been no increase in head circumference and computerised tomography showed widespread cerebral atrophy with bilateral subdural effusions. Plasma protein electrophoresis showed that the α -1 band was greatly reduced. The concentration of α -1 antitrypsin was 0.5 g/l and phenotype homozygous Pi Z (father: 0.9 g/l, MZ; mother: 1.2 g/l, MZ; adult reference interval 1.8–3.0 g/l). A liver biopsy specimen showed extensive fibrosis with PAS-positive diastase-resistant granules in the periportal zones, and electron microscopy also showed the typical appearance of α -1-antitrypsin deficiency.

We think that the bleeding disorder was caused by the α -1-antitrypsin deficiency and as a result she suffered an intracranial haemorrhage with extension into the sub-arachnoid space. We confirm the conclusions of Hope *et al.*¹ that α -1-antitrypsin deficiency may present as a bleeding diathesis. Early diagnosis and treatment are needed to prevent devastating sequelae.

Reference

- 1 Hope P L, Hall M A, Millward-Sadler G H, Normand I C S. Alpha-1-antitrypsin deficiency presenting

as a bleeding diathesis in the newborn. *Arch Dis Child* 1982; **57**: 68–70.

H R JENKINS, J V LEONARD, AND J D S KAY
*The Hospital for Sick Children,
Gt Ormond Street,
London WC1N 3JH*

R W POOL, J A SILLS, AND D M ISHERWOOD
*Alder Hey Children's Hospital,
Eaton Road, Liverpool*

This correspondence is now closed: *Editor*.

New sign of tracheo-oesophageal fistula?

Sir,

The classical presentation of oesophageal atresia with tracheo-oesophageal fistula is the 'bubbly' baby who has difficulty swallowing his or her own saliva and whose respiration becomes noisy.

I offer a new sign of tracheo-oesophageal fistula, tentative because in clinical medicine new clinical signs are often signs rediscovered, and also because it is based on observations in a single baby. He was born by elective caesarean section at 40 weeks' gestation. There were no clinical signs of polyhydramnios and no excess of amniotic fluid noted when the uterus was incised. Resuscitation was routine although the baby became secondarily apnoeic at 5 minutes and had to be aspirated under direct vision, when a small amount of viscid mucus was obtained. One hour later a similar thing happened but the baby was not 'bubbly'.

An attempt to pass a nasogastric tube failed and x-ray films showed the curled up tube in the upper third of the oesophagus. Auscultation of the chest was equal on both sides but the striking feature was that on auscultation of the abdomen, especially the upper half, breath sounds were as easily heard as over the chest, although they had a slightly resonant or amphoric quality. On reflection this would not be surprising in the common type of tracheo-oesophageal fistula where the middle portion of the oesophagus is atretic and the lower third moiety communicates with the trachea. In the baby we described this communication was wide and joined with the trachea at its bifurcation.

I should like to know whether other workers have either noticed this sign or have read about it.

D M LEWIS
*Department of Paediatrics,
Bronglais General Hospital,
Aberystwyth SY23 1ER*